

## Buschke scleredema: A case report

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**Abstract** Scleredema is a rare skin disease “progressive hardening of the skin” as the cardinal feature. Here we report a case in a four year old girl with this rare condition.

**Key words**

Scleredema of Buschke, scleredema.

### Introduction

Scleredema of Buschke is a fibromucinous connective tissue disease characterized by massive deposition of mucin in collagen bundles of skin eventually causing hardening of skin<sup>1,2</sup>. Generally it occur post infectious<sup>3,4</sup> e.g. pharyngitis. It also has significant association with diabetes. Some cases of scleredema are reported in patients of multiple myeloma, HIV, rheumatoid arthritis and Steven-Johnson syndrome<sup>5,6</sup>. Our patient presented to us with this rare manifestation after upper respiratory tract infection and the diagnosis was confirmed by histopathology.

### Case report

A four year old girl presented in OPD of dermatology department at KMSMC, Sialkot, accompanied by her father. Her chief complaint was hardening of the whole body skin causing difficulty in mouth opening and flexion of the limbs. According to her father all these symptoms appeared within a span of two weeks. Otherwise, the girl was healthy and had no medical issues.

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On examination of the skin it was found to be hard, indurated and pale, particularly upper limb skin was hard as compared to lower limb.

Routine blood examination, urine analysis along with chest X-ray and ultrasound abdomen had normal findings.

Treatment was started following report of the punch biopsy which shows thickened dermis and thickened collagen.

We gave patients a five day course of antibiotics and topical steroids which gave satisfactory results and the symptoms improved markedly.

### Discussion

Scleredema of Buschke occurs in all age groups although 50% of cases are reported in patients younger than 20 years old. The ratio of female affected with this disease is twice than the male ratio (2:1).

Disease presents with acute progressive hardening of the skin leading to restriction of body movements, dysphagia, dysarthria and difficulty in closing eyes<sup>4</sup>.

Three forms of scleredema of Buschke are commonly seen.



**Figure 1** Hard, indurated, pale and progressive hardening of the skin.



**Figure 2** Thickened dermis forming gaps between collagen bundles

**Type 1** post- infection. This type is self-limited and generally resolves without treatment in four to six weeks.

**Type 2** is associated mainly with multiple myeloma. It requires adequate management as spontaneous remission is less likely

**Type 3** known as Scleredema diabeticorum is associated with diabetes. Control of glucose level has no relation with the improvement of disease. The course of this type is refractory and more consistent as compared to rest of the types.

The histopathology of skin sample shows thickened dermis because of excessive deposition of mucopolysaccharides in the dermis. Toluidine blue stain is preferably used to attain adequate findings of the histopathology.

The post-infectious scleredema is generally a self-limited illness which requires no treatment but in some cases antibiotics and steroids have shown better outcome of the condition<sup>7</sup>.

In progressive and acute disease IVIG is recommended. Also UVA -1 phototherapy in combination with methotrexate or colchicines have given the desired results<sup>8,9</sup>.

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